Journal of Medical Ultrasound (2016) 24, 25—28

Chinese Taipei Society of Ultrasound in Medicine

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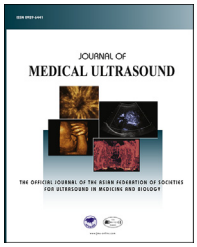

TECHNICAL NOTE

Ultrasonic Contrast Portography for Demonstration of Intrahepatic Porto-systemic Shunts

CME Credits

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Received 15 November 2015; accepted 7 December 2015
Available online 29 January 2016

KEYWORDS
color Doppler ultrasound,
contrast-enhanced ultrasound,
liver,
shunts,
ultrasonic contrast agent,
veins

Abstract Spontaneous intrahepatic porto-systemic shunts (IHPSS) can be disclosed with ultrasound (US) and color Doppler ultrasound (CDU). However, direct evidence of the shunt on US or CDU may not be convincing. In this report we demonstrate the presence of IHPSS by ultrasonic contrast portography with intravenous injection of microbubble-based contrast agent (MBCA). With this technique, the MBCA was depicted to enter the hepatic vein through the shunt, and then flowed into the inferior vena cava.

Introduction

Spontaneous shunt between portal veins and hepatic portal and hepatic veins, an intrahepatic vascular malformation, is a rare disease entity [1—7]. Early diagnosis is important to prevent hepatic encephalopathy and hematemsis. Fewer than 50 cases of spontaneous intrahepatic porto-systemic shunts (IHPSS) have been reported in the literature [6—11]. IHPSS may be disclosed with ultrasound (US) or color Doppler ultrasound (CDU) [10—12]. However, direct evidence of IHPSS on US or CDU may not be convincing. In

Conflicts of interest: All authors have no conflicts of interest to declare.

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http://dx.doi.org/10.1016/j.jmu.2015.12.001

this report we demonstrate the IHPSS by using ultrasonic contrast portography with intravenous injection of microbubble contrast agent (MBCA).

Techniques

A suspension of galactose microparticles in distilled water (Levovist; Schering, Berlin, Germany) was used as an MBCA. The microbubbles formed after vigorous shaking of the microparticle suspension for 10 seconds are estimated to have an average diameter of 2—8 μm. A total of 2.5 g of levovist in a concentration of 300 mg/Ml (8 mL) was injected manually at a speed of 1 mL/s through a 20-gauge cannula inserted in the antecubital vein, and was flushed with an additional 10 mL of normal saline. A US study was performed using a Logiq 9 scanner (GE Medical System, Milwaukee, WI, USA) equipped with coded harmonic function. A 24-MHz curved linear array broadband transducer was used. The mechanical index was set at 0.60.8. Before the contrast study, a routine fundamental hepatic sonography was performed to demonstrate the location with suspected abnormal vasculature. The best scanning plane was chosen, and then the coded harmonic US function was switched on. Ten seconds after injection of Levovist suspension the patient was instructed to hold his breath so that the blood flows in the vessels of the porta hepatics enhanced by the MBCA could be continuously monitored and the suspected abnormality could be traced. The whole course of the US contrast study was recorded with a compact disk recorder.

Illustration of a case

A 70-year-old man was admitted with the chief complaints of abdominal fullness and two episodes of passage of tarry stool in the past month. The past history was unremarkable except for occasional nose bleeds. On physical examination, he had anemic conjunctivae and a few telangiectasis on the tongue, lips, nasal septum, and the turbinates bilaterally. He had a regular pulse with a Grade 2/6 precordial systolic murmur on auscultation. The vertical span of the right hepatic lobe was ~15 cm on percussion, indicating mild hepatomegaly. The other parts of abdomen were otherwise normal. The laboratory values were within normal range except for a minimally elevated alkaline phosphotase (220 IU/L) and alanine aminotransferase (52 IU/L). To investigate the cause of hepatomegaly, a US study of the upper abdomen was undertaken, which disclosed dilated common hepatic artery (maximal diameter: 8.5 mm). Multiple tortuous tubular structures representing vascular channels in the right hepatic lobe were noticed. By careful US tracing and color Doppler US evaluation of the right lobe vessels, communication between the right portal branch and the right hepatic vein through these vascular channels was suspected (Figure 1). Based on the clinical manifestations and US findings of the liver, a diagnosis of Osber—Weber—Rendu disease (OWRD) was made. The liver pathology was attributed to IHPSS. To verify the existence of shunting, intravenous ultrasonic contrast portography was done. Cloud-like contrast-enhanced blood flow was demonstrated in the right portal

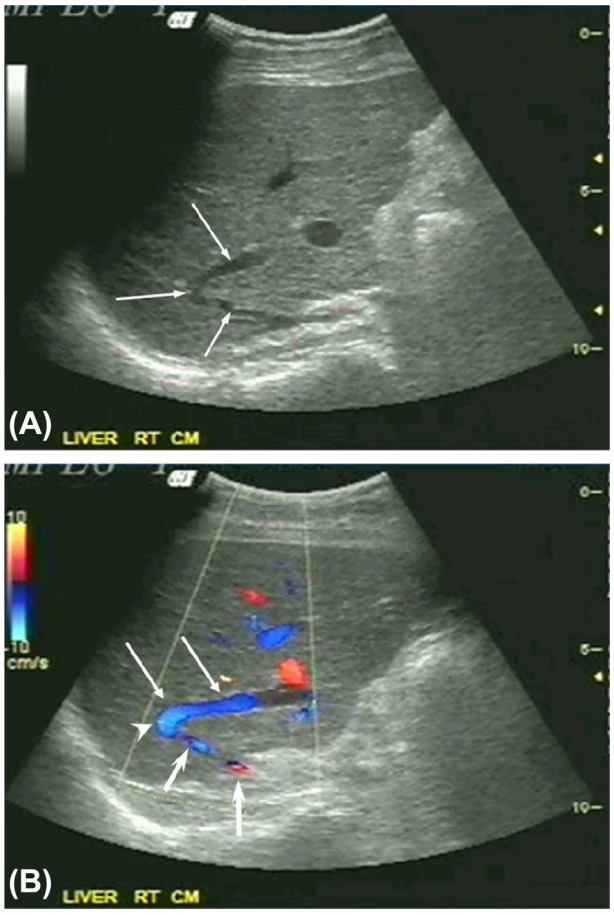

Figure 1 Gray-scale and color Doppler US studies of the liver. (A) Gray-scale US of the right hepatic lobe (intercostal scan) demonstrates a venous structure which is suspected to run in a curved course (arrows), and connection between a portal branch and a hepatic vein is possible; (B) color Doppler US depicts a possible communication (arrowhead) between a branch of the right portal vein (small arrows) and the right hepatic vein (large arrows). However, the continuity of the vessels cannot be well portrayed. US = ultrasound.

branch. Via the communicating collateral venous channels, the enhanced blood ran toward the right hepatic vein and then to the inferior vena cava (Figure 2). A diagnosis of hepatic angiodysplasia with IHPSS was confirmed on the basis of intravenous ultrasonic contrast portographic findings. The patient was therefore treated conservatively for his gastrointestinal bleeding, and has been doing well since then for 3 years.

Discussion

The cause of IHPSS is still unclear. It has been contributed to injury, congenital malformations, or collaterals as a result of portal hypertension such as in cirrhosis of the liver and Budd—Chiari syndrome [10—13]. Patients with SPVS can be asymptomatic and their vascular abnormalies are incidentally found during abdominal imaging. However, several clinical manifestations can be present at the time of diagnosis, including hematemesis, ascites, abnormal behavior,

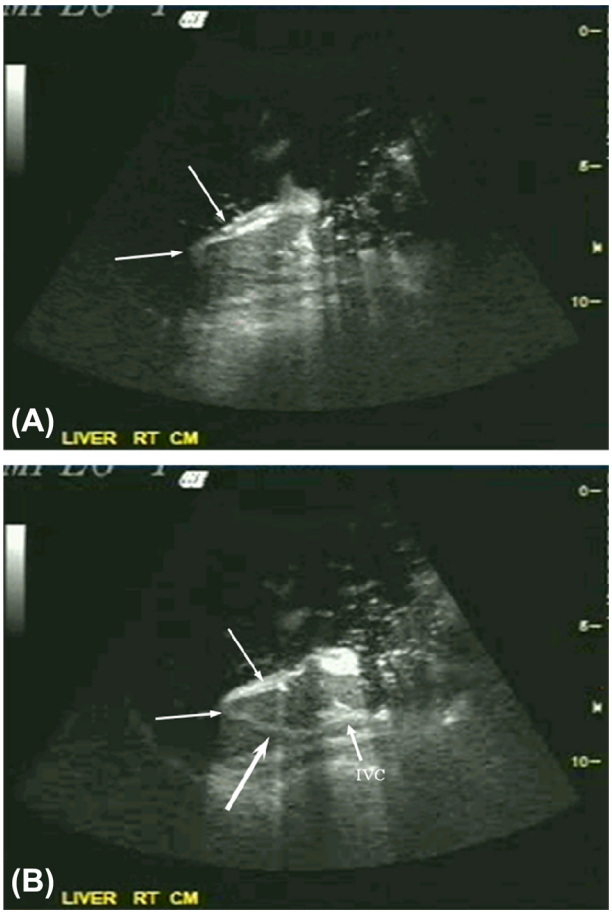

Figure 2 Contrast-enhanced US after intravenous MBCA injection. (A) 16 seconds after injection: the right portal branch (arrows) is enhanced; (B) 18 seconds after injection: the hepatic vein (large arrow) and the inferior vena cava (IVC) lumens are enhanced by the microbubbles and become echogenic. The continuity of the vessels is well shown. MBCA = microbubble-based contrast agent.

loss of consciousness, and flapping tremor. Some of the latter symptoms are related to hepatic coma [14—16]. The most commonly reported cases with symptoms usually have a single large tube of constant diameter that connects the right portal vein to the inferior vena cava [6]. This is the most common type and has been classified as Type I IHPSS. There are some other morphological types of IHPSS. The second type is a localized peripheral shunt in which single or multiple communications are found between peripheral branches of the portal and hepatic veins in one segment [15]. The third type presents as an aneuysmal fistula between the communicated portal and hepatic veins [17]. The fourth type has multiple communications between peripheral portal and hepatic veins diffusely in both lobes of the liver [6]. Our cases correspond to the first type. Recognition of this sort of vascular malformation is therefore of clinical importance because there could be significant clinical symptoms developed during the patient's lifetime.

The hepatic vasculature is best evaluated with conventional or substraction iodinated contrast angiography. Since the development of cross-sectional imaging modalities such as computed tomography (CT), US, and magnetic resonance imaging (MRI), noninvasive demonstration of the hepatic vasculature has been made possible. These techniques, also referred to as CT angiography, MR angiography, and ultrasonic angiography in their state-of-the-art functions have been applied in clinical studies of the visceral vessels [18—23]. Among these, US in conjunction with various Doppler technologies is the most convenient method in the evaluation of hepatic vasculature [23,24]. Power Doppler US may provide more information regarding the shunts between hepatic arteries and veins, and better demonstration of dilated and tortuous hepatic arteries [5]. In the present case, the dilated communicating venous channels between the portal veins and the hepatic vein can be traced with real-time US and demonstrated clearly with power Doppler US. In patients with IHPSS, the dilated feeding hepatic artery in the hepato-duodenal ligament and porta hepatis may mimic a dilated biliary duct sonographically. Similarly, the dilated communicating venous channel can be also mistaken as dilated intrahepatic bile ducts, the so-called too-many-tubes sign. Color Doppler and/or power Doppler US can portray color flow signals in the aforementioned dilated artery or venous channels Doppler. Power Doppler is particularly good for delineation of the vascular course in a more continuous fashion [5]. However, in some vascular channels when the vessels are extremely tortuous or intervening, color Doppler and power Doppler US may have difficulty in demonstrating the relationship of the closely arranged vessels. Iodinated contrast venography is the gold standard in this instance. With the recent advent of ultrasonic contrast agents, it is possible to perform contrast-enhanced ultrasonography (CEUS) via intravenous route and to observe the echo-enhancing effect in the target organs or pathologies [25—27]. CEUS can also be used to evaluate the transjugular intrahepatic portosystemic shunt (TIPS) and its complication [28]. Levovist, a galatose-based contrast agent, can enter the portal vein via the systemic circulation ~15—20 seconds after intravenous administration of the levovist suspension solution. The vasculature of hepatic tumors may be significantly enhanced with intravenous MBCA [26,27]. In this paper, CEUS by injection of MBCA into the autecubital vein demonstrated MBCA flowed into the right portal branch, the shunts, and the drainage hepatic veins and IVC were all enhanced with echogenic microbubbles. The direct demonstration of the communication enhanced the diagnostic confidence of IHPSS for some patients in whom noninvasive imaging may not be conclusive. This minimally invasive method can be safely monitored under real-time US [29].

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